Third Ventricle Immature Teratoma: A Case Report

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We present the case of a 1-year-old girl with persistent vomiting who was found to have a disproportionately large immature teratoma in the third ventricle. Magnetic resonance imaging demonstrated a heterogeneous intracranial mass in the third ventricle, with a compressed left cerebral hemisphere and hydrocephalus. Bifrontal craniotomy via a transchiasmatic approach achieved total resection of the tumor. No significant neurologic deficit was seen except for double vision and disturbances in eye movement. Third ventricle immature teratoma, although extremely rare, may present as a suprasellar mass lesion and hydrocephalus. The prognosis after gross total resection is favorable; however, radiotherapy is usually postponed until 36 to 48 months of age in order to avoid complications such as stunted growth, endocrine disturbances and neuropsychologic problems.

Key Words: immature teratoma, transchiasmatic approach
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Intracranial germ-cell tumors are rare and account for 0.3% to 2% of all pediatric intracranial neoplasms [1]. All except pure germinoma and mature teratoma are malignant. Although most intracranial germ-cell tumors arise in the midline, the pineal gland is the most common site of origin. Occasional examples are situated laterally in the basal ganglia or thalamus. This third ventricle immature teratoma is the first documented case described in the English literature.

Case Presentation

A 1-year-old girl presented with a fever of 38.5°C with vomiting and poor appetite for 6 days and scarlatiniform rash over the trunk. Under the initial impression of acute gastritis and roseola, she was admitted and treated conservatively. However, her clinical condition did not improve. There was frequent vomiting, disturbed consciousness, poor appetite and limited neck motion accompanied by visual field defect and blurred vision. The head circumference was within the normal range, but visual evoked potential revealed no wave response bilaterally. After epidural drainage, cerebrospinal fluid studies revealed pleocytosis with a high neutrophil/lymphocyte ratio (5/95). Magnetic resonance imaging (MRI) revealed a well-defined encapsulated mass lesion measuring 4.0 × 4.0 × 3.5 cm with a cystic and solid component, as well as hydrocephalus producing dilated ventricles (Figure 1). Serum and cerebrospinal fluid α-fetoprotein (AFP) concentrations were significantly elevated at 13,740 and 574 ng/mL, respectively. Serum β-human chorionic gonadotropin (β-hCG) concentration was mildly elevated at 17.72 ng/mL. Serum carcinoembryonic antigen (CEA) and cerebrospinal fluid β-hCG were both within normal ranges.

Considering the patient's clinical condition and progressive decline, we decided that surgical intervention was

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Figure 1. *(A) Contrast-enhanced computerized tomography image showing a large third ventricle tumor mass with marked dilatation of the lateral ventricles. (B) Sagittal gradient echo (GE) T2-weighted magnetic resonance (MR) image of the brain (TR/TE = 400/15) with gadolinium enhancement prior to surgical intervention reveals cystic and enhanced solid components and marked calcification of the suprasellar area. Note the marked dilatation of the ventricles. (C) Coronal GE T2-weighted MR image of the brain (TR/TE = 400/15) with gadolinium enhancement prior to surgical intervention.*

required to remove the tumor. The tumor was approached via a midline bifrontal craniotomy and was totally excised piecemeal. Intraoperatively, the dura was bulging and bluish. The optic chiasma was pale and swollen, which is compatible with the MR image of compression by the tumor. The tumor was exposed via a left paramedial incision in the optic chiasma. The tumor and its surrounding capsule were nearly totally removed.

The tumor was relatively hypovascular, with a cystic formation containing hair particles and gelatinous material. Histopathology revealed an immature teratoma composed of a variety of immature tissues derived from three embryonic germ layers, but no elements of yolk sac tumor or embryonal carcinoma. In addition to the immature mesenchymal tissue, there were considerable amounts of cuboidal or columnar epithelium with immature goblet cells, islands of immature cartilage, and immature adipose tissue. The primitive tissues showed developing neuroectodermal differentiation with rosette formation of neuroepithelial cells (Figure 2A–C). Strong positive staining for AFP was present in some nested hepatoid tissues (Figure 2D), and immunoassay for glial fibrillary acidic protein showed large areas of neuroectodermal tissue.

Hydrocephalus improved soon after tumor removal. General activities improved, but poor appetite and lethargy were noted. Postoperative MRI revealed bifrontal epidural hematoma and extensive bilateral frontotemporoparietal effusion with some mass effect (Figure 3). There was no evidence of residual tumor in the third ventricle. Two days after surgery, serum AFP and β-hCG decreased to 57 and 0.3 ng/mL, respectively, and cerebrospinal fluid AFP decreased to 178 ng/mL. The serum cortisol concentration was significantly decreased and cortisol acetate (0.7 mg/kg/day) was prescribed. Epidural drainage was removed 1 week after the operation. Postoperative 1-year follow-up revealed no evidence of recurrence, and the patient was in a stable condition at the time of writing.

Discussion

Congenital intracranial immature teratoma is rare. It accounts for 0.3% to 0.7% of all intracranial tumors, and up to 0.3% to 2% of childhood intracranial neoplasms [2]. Takaku et al found a high incidence of these lesions (> 50%) in children aged less than 2 months [3]. The World Health Organization classification of intracranial teratoma considers three distinct histologic variants, mature, immature and malignant, making up 0.3% of all intracranial neoplasms [2]. Intracranial teratoma is thought to originate from either somatic or ectopic germ cells. The components are often immature, and include primitive mesenchymal tissue and primitive neuroectodermal structures, occasionally in true rosette formation [1,4]. The tumor is located supratentorially more often in infants than in older children [5].

Differential diagnoses include astrocytoma, ependymoma, primitive neuroectodermal tumor, other germ cell tumors and choroid plexus papilloma [3]. The most common clinical presentations of immature teratoma are macrocrania, hydrocephalus and a bulging fontanel. Signs of increased intracranial pressure, such as papilledema and nuchal rigidity, are less frequent because of decompression through the

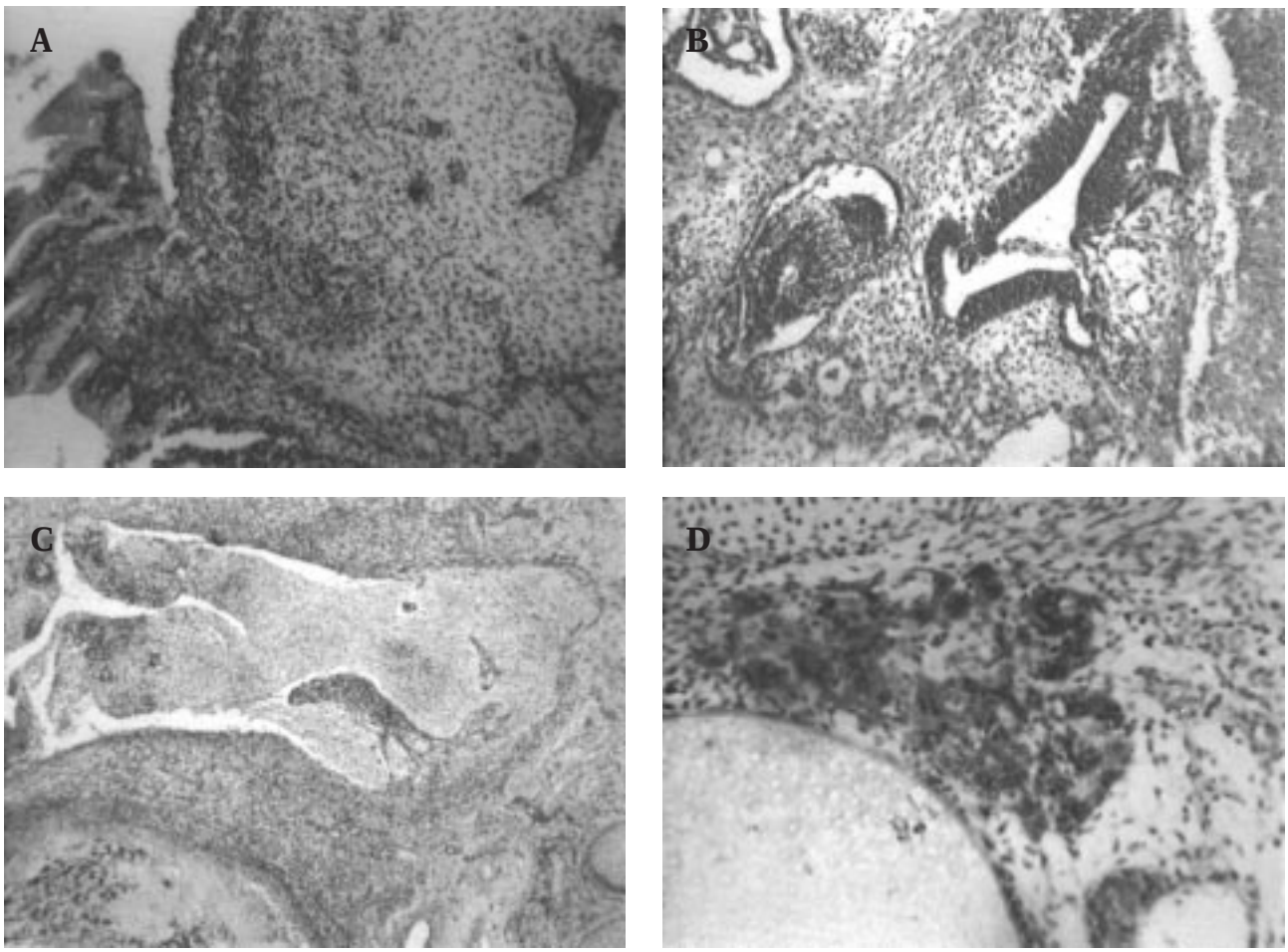

Figure 2. *Histopathology of the tumor specimen. (A) Islands of immature cartilage, ossifications, primitive intestinal epithelium and skin adnexa (hematoxylin & eosin, ×100). (B) Developing neuroectodermal tissues consisting of primitive neuroepithelial cells with pseudostratified tubular and rosette formations. Note the presence of primitive mesenchymal tissue (hematoxylin & eosin, ×400). (C) Primitive glial tissue and immature ossification and cartilage formation (hematoxylin & eosin, ×100). (D) Hepatoid tissues consist of hepatoid cells arranged in solid nests; immunohistochemical studies reveal a strong positive staining reaction for α-fetoprotein (α-fetoprotein, ×400).*

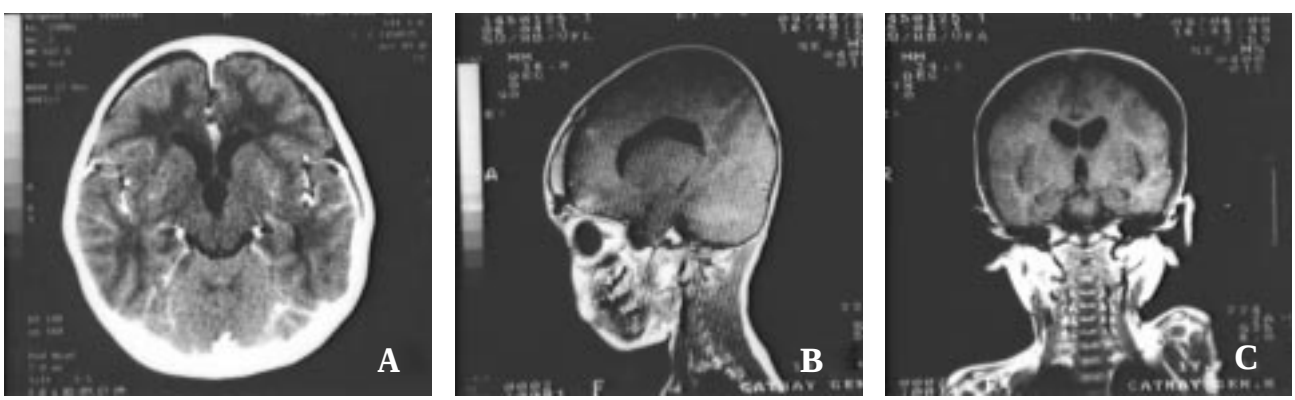

Figure 3. *(A) Contrast-enhanced computerized tomography image showing a subdural effusion in the bilateral frontoparietal area; there was no evidence of tumor recurrence. (B) Sagittal gradient echo (GE) T2-weighted magnetic resonance (MR) image of the brain (TR/TE = 400/15) with gadolinium enhancement after surgical intervention reveals mild hemorrhage over the medial bifrontal region. (C) Coronal GE T2-weighted MR image of the brain (TR/TE = 400/15) with gadolinium enhancement after surgical intervention.*

cranial sutures. Other findings include paresis, cranial nerve deficit, convergence nystagmus, seizures, vomiting and lethargy [6]. Visual disturbances, including diplopia, decreases in visual acuity, ptosis and blindness, have been reported in 75% of patients. Diabetes insipidus is the most common endocrine manifestation [1,6]. Other endocrine disturbances include growth retardation, abnormal menses, precocious puberty and galactorrhea.

MRI is the most valuable complementary tool when prenatal ultrasonography is incomplete, doubtful and limited [2,7,8]. It is most useful in making the diagnosis of fetal central nervous system anomalies and fetal death because lack of fetal movement and lesions are clear enough to be detected. Immature teratoma is hypointense on T1-weighted image and slightly hyperintense on T2-weighted image. Both show the presence of perifocal edema. Computerized tomography (CT) is superior to MRI in the detection of calcification, but MRI, through its multiple image planes and absence of bone artifacts, better delineates the extent of the tumor, especially within the posterior fossa [8, 9]. An isodense or low-density mass mixed with cystic and solid components may be found on plain CT scan. Marked perifocal edema may also be observed [8].

The most frequent histologic findings are fetal-appearing tissue of the type seen in "products of conception" [5]. In this melange, some developing neuroectodermal structures are often seen, such as embryonal medullary neuroepithelium, retina and choroid plexus. Common mesenchymal elements include cellular or myxoid stroma. Islands of cartilage, occasionally undergoing ossification, and intersecting bands of striated muscle are usual [1]. Immunohistochemically, immature teratomas express reactivity appropriate for epithelial and mesenchymal differentiation. The epithelial component may be positive for CEA, and scattered syncytiotrophoblasts are predictably positive for β-hCG [1]. In immunohistochemical studies, AFP is detected in the cytoplasm of gastrointestinal tissue and primitive neuroepithelial elements, and is rarely seen in immature teratoma [1,10]. However, Nakashima et al reported that hepatoid nests were observed in 11% of immature teratoma [11]. In addition, Toki et al reported a case of immature retroperitoneal teratoma and demonstrated, by immunohistochemical studies, that the AFP in the serum was derived from hepatoid cells in the tumor [12]. In our opinion, AFP elevation in serum and cerebrospinal fluid does not indicate a poor prognosis in immature teratoma.

The prognosis for immature teratoma is extremely poor because of the rapid, invasive growth of the tumor and the destruction of normal brain structures [2]. Treatment includes radical surgical resection followed by radiotherapy and/or chemotherapy [2,13,14]. The extent of surgical resection is an important prognostic factor. Herder and Ventureyra reported recurrence 4 months after successful surgery [15]. The operative mortality for intracranial tumor in the first year of life is 20%. However, immature teratomas may show widely varying responses to adjuvant therapy, leading to striking differences in disease-free survival after the same therapeutic regimens [16]. The overall 5-year survival rate was 18% for malignant teratomas in a series reported by Ferreira et al [7]. If the patient survives, radiotherapy is usually postponed until 36 to 48 months of age in order to avoid complications such as stunted growth, endocrine disturbances and neuropsychologic problems [3,16,17].

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第三腦室未成熟畸胎瘤一病例報告

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第三腦室未成熟畸胎瘤在臨床上是極為少見的病例，通常此類的病人往往表現出水腦或是顱內腫瘤引發之顱內壓升高等等症狀。我們提出一十個月大的女嬰，出現有嚴重的頭痛、嘔吐等顱內壓升高的問題，在核磁共振檢驗之下，發現有第三腦室腫瘤合併有水腦的問題，因此緊急採取手術摘除腫瘤及減壓，術後病人症狀得到了顯著的改善。我們將回溯文獻有關於未成熟畸胎瘤的臨床症狀、手術治療及預後，並討論手術、放射治療和其預後的關聯。

關鍵詞：未成熟畸胎瘤

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